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Neuralgic Amyotrophy, also known as Parsonage-Turner Syndrome (PTS), is a clinically diagnosed and largely under-recognized inflammatory disorder which primarily affects the brachial plexus. Correct diagnosis is often delayed and the disease causes significant patient morbidity due to extreme pain, paresis, and often sensory abnormalities of the upper extremity. Although the pathophysiology is not completely understood, it is classically preceded by an identifiable inciting event and complete recovery may take months to years.

A Case of Parsonage-Turner Syndrome

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Abstract

Neuralgic Amyotrophy, also known as Parsonage-Turner Syndrome (PTS), is a clinically diagnosed and largely under-recognized inflammatory disorder which primarily affects the brachial plexus. Correct diagnosis is often delayed and the disease causes significant patient morbidity due to extreme pain, paresis, and often sensory abnormalities of the upper extremity. Although the pathophysiology is not completely understood, it is classically preceded by an identifiable inciting event and complete recovery may take months to years.

Introduction

Shoulder pain is the third most common presenting musculoskeletal complaint in primary care settings.[1] However, due to the shoulder joint's complex anatomy, accurate diagnosis and management for uncommon illnesses is often delayed. These delays can lead to worse outcomes and increased morbidity for the patient. One under-recognized etiology of shoulder pain is Neuralgic Amyotrophy, also known as Parsonage-Turner Syndrome (PTS). PTS is a clinically diagnosed inflammatory condition of the brachial plexus characterized by acute onset shoulder pain, muscle paresis, and varying amounts of sensory abnormalities, which often occurs after an identifiable inciting event. Although once considered a rare disease, more recent studies indicate an annual incidence of closer to 1:1000.[2] Therefore, primary care physicians must maintain a high index of suspicion for this disorder when evaluating patients presenting with shoulder pain. We present a case of PTS which had an excellent outcome for which we obtained consent to use this case for educational purposes.

Case

A 55 year old, right hand dominant male was referred to Sports Medicine Clinic for evaluation of abrupt onset right shoulder pain and weakness. The patient awoke with symptoms two days prior and was evaluated by his primary care provider (PCP). He described his pain as diffuse and dull with additional sharp, stabbing pain along the cervical spine and medial scapula. He also reported that he could no longer raise his right arm, although he denied paresthesias. There was no known mechanism of injury. Review of systems was notable for "flu-like" symptoms including fever, cough, and myalgias starting approximately forty-eight hours prior to onset of symptoms. There was no significant past medical, surgical, or social history.

On physical exam, the patient demonstrated superior right shoulder carriage. No skin findings were noted, and the shoulder was non-tender to palpation. He had preserved, and pain free, passive range of motion in all planes. Active motion was severely limited due to 1/5 strength with shoulder flexion, resisted external rotation, as well as elbow flexion and forearm supination. Grip strength was preserved. He also had diminished biceps, triceps, and brachioradialis reflexes without noted upper extremity sensory deficits. The cervical spine examination was unremarkable.

A diagnosis of PTS was considered, although the differential also included cervical radiculopathy, cerebrovascular accident, Todd's paralysis, and spinal infection including vector borne illnesses. A negative chest radiograph taken at initial presentation was reassuring against Pancoast tumor.

The case was reviewed with Neurology who recommended hospital admission for further management. A viral panel was positive for Influenza A. Chest and shoulder radiographs, as well as magnetic resonance imaging (MRI) of brain, neck, and shoulder were unremarkable. Lumbar puncture was negative for infection. Based upon this clinical picture, the diagnosis of PTS was made and the patient was started on IV Methylprednisolone. The patient began demonstrating clinical improvement and by day three the patient was pain free and had notable improvements in strength (3-4/5 in all directions) and normalization of reflexes.

The patient was discharged from the hospital on a 60mg Prednisone taper for 10 days and continued close outpatient follow-up in Sports Medicine Clinic while also completing physical therapy regularly. At four month follow-up the patient's shoulder exam had normalized and he had a full return of baseline strength without pain or work/lifestyle limitations.

Discussion

PTS is predominantly a clinical diagnosis with the classic presentation of abrupt onset, intense, unilateral shoulder girdle pain, often nocturnal at onset, with subsequent and progressive muscle weakness and varying levels of sensory abnormalities. Although the pathophysiology is not completely understood, it is likely immune-mediated as there is a high reported incidence (up to 50%) of preceding infectious etiologies.[3,4] Other common triggers include exercise, surgery, vaccination, and pregnancy.[2-6] A pathognomonic characteristic of PTS is that pain, paresis, and sensory symptoms classically do not affect the same nerve distributions and that passive range of motion is preserved. These characteristics are especially important when differentiating PTS from cervical radiculopathy and glenohumeral bursitis, which are the most common incorrect diagnosis initially given.[3,5]

The importance of recognizing these clinical features becomes even more critical due to often nonspecific or unremarkable ancillary testing. Electromyography (EMG) and findings on MRI may demonstrate patterns of muscle denervation, but these findings are not diagnostic or specific to PTS.[5,7] There is growing interest in the use of ultrasound detection of morphologic changes to affected nerves, but further studies are needed before generalization.[5,7] Unfortunately, due to provider unfamiliarity and lack of specific findings on ancillary testing, diagnosis is frequently delayed for several months.[3,5] The mainstay for treatment of PTS involves early initiation of corticosteroids and prolonged courses of PT/OT focusing on range of motion and function. Strength training should not be a priority as overloading muscles during reinnervation can lead to increased muscle strain and prolonged pain/recovery.[4-6] Even with proper management, full recovery may take months to even years.[4-6]

Conclusion

Our case illustrates the importance of having a broad differential and completing a thorough ROS in patients with shoulder pain. PTS is an under recognized cause of acute onset unilateral shoulder pain and paresis which is often preceded by an inciting event. A high index of suspicion should be maintained as prompt identification and management may decrease the high patient morbidity associated with this diagnosis. ■

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